

ERNST HANHART (1891-1973) AND THE SYNDROME WHICH BEARS HIS NAME

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Richner-Hanhart syndrome (RHS) is a rare autosomal recessive disease associated with high serum tyrosine levels. RHS is caused by the deficiency of tyrosine aminotransferase enzyme (TAT) [1-5]. It is listed in Online Mendelian Inheritance of Man as (tyrosinemia type II) (OMIM #276600). It is a disorder of amino acid metabolism which characterized by ocular changes, painful palmoplantar hyperkeratosis, and mental retardation. Serum tyrosine increases resulting in the deposition of tyrosine crystals in the cornea and in corneal inflammation [5].

It is mapped to gene 16q22.2. Patient with RHS may present with complaints of bilateral photophobia and tearing, which started during the infancy period. RHS should be suspected in patients demonstrating dermatologic signs, especially palmoplantar keratosis, associated with bilateral pseudodendritic corneal lesions unresponsive to antiviral therapy [1].

Patients are often misdiagnosed as having herpes simplex keratitis. However, serum and urine tyrosine levels confirm, usually, the diagnosis. A low tyrosine and phenylalanine diet permitted good control of the disease with a complete resolution of the oculo-cutaneous symptoms in a month. The importance of an early diagnosis of this syndrome to avoid the risk of mental retardation was emphasized [3].

This syndrome is reported first by Richner [6] in 1938 and Hanhart [7] in 1947.

Dr. Hermann Richner, was a Swiss dermatologist, born September 6, 1908, Zürich [8].

Ernst Hanhart (1891-1973), was a Swiss internist and human geneticist [9] (Fig. 1). After qualifying in medicine from the University of Zurich in 1916. He worked under professors Otto Nägeli (1871-1938) and Wilhelm Löffler (1887-1972). Hanhart became interested in human genetics and became a specialist in hereditary disorders [9].

He was appointed professor at the University of Zurich in 1942 and was a founding member of the Swiss Society of Genetics [9].

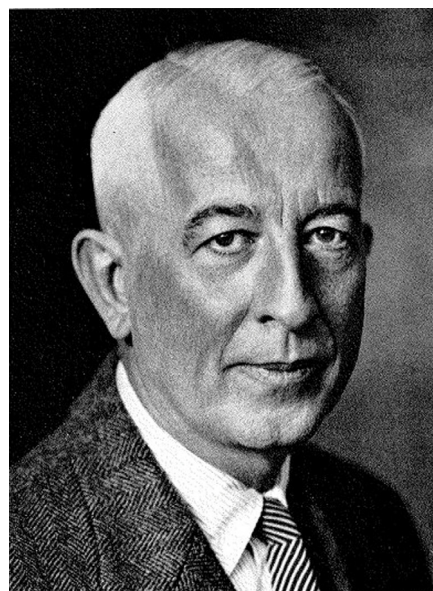
*Ernst Hanhart*

Figure 1. Ernst Hanhart (1891-1973).
Courtesy of Prof. Dr. med. Anita Rauch University of Zurich, Institute of Medical Genetics.

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